Hyperkinetic Movements in Children:
Differential Diagnosis, Evaluation, and Treatment

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LESSONS LEARNED AT THE INTERFACE OF MEDICINE AND PSYCHIATRY

The Psychiatric Consultation Service at Massachusetts General Hospital sees medical and surgical inpatients with comorbid psychiatric symptoms and conditions. During their twice-weekly rounds, Dr Stern and other members of the Consultation Service discuss diagnosis and management of hospitalized patients with complex medical or surgical problems who also demonstrate psychiatric symptoms or conditions. These discussions have given rise to rounds reports that will prove useful for clinicians practicing at the interface of medicine and psychiatry.

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Hyperkinetic movement disorders can appear straightforward; however, they can also be complex and rare, making misdiagnosis and ineffective management more likely.[1] Therefore, we seek to provide primary care providers with a foundation for understanding pediatric hyperkinetic movement disorders, including tips on creation of a differential diagnosis, assessment of risk factors, and guidance on management strategies.

Case Vignette 1

A 4-year-old boy was brought to an outpatient clinic for an evaluation of extra movements that he had since the age of 1 year. He met all of his developmental milestones on time. His movements began with hand flapping, and they evolved over several years into more complex movements that included walking in circles and repeatedly jumping up and down. The movements occurred when he was excited, nervous, or stressed. Since their onset, the movements had not changed in character. The movements did not cause psychosocial, educational, or physical impairments. There was no family history of a movement disorder. The child had a normal neurologic examination, with only a brief instance of jumping up and down along with hand flapping observed during the clinical visit. His parents preferred that he receive conservative management.

Case Vignette 2

A 7-year-old boy presented to an outpatient clinic for a second opinion regarding extra movements that were previously diagnosed as tics. He had been diagnosed with a developmental coordination disorder and a phonologic disorder. Around the age of 3 years, he developed stereotyped movements of his head (moving to the right or left), with extension of his arms and legs. These movements occurred with anxiety and stress. They had not changed in character since their onset. There was a family history of Tourette syndrome (TS). The movements had not caused educational, psychosocial, or physical impairment. However, his parents were concerned about the potential for future bullying/teasing. On examination, he had choreiform movements, signs of motor impersistence (which is the inability to sustain a movement such as "milk-maid's grip" or "darting tongue"[2]), and hypotonia. The stereotyped and choreiform movements did not bother the child; his parents wanted him to receive conservative management.

Case Vignette 3

A 12-year-old boy presented to an outpatient clinic for evaluation of tics. He developed tics at the age of 5.5 years, with movements of his jaw and neck and hair twisting. He also made throat-clearing and clicking sounds. His motor and vocal tics waxed and waned, and they changed in frequency, severity, and presentation. When he was younger, he had predictable extra movements that included hand flapping and

Clinical Points

- Identification of the type of movement disorder is crucial.
- Clues in the history (including family history) and examination can guide creation of a differential diagnosis and the diagnostic workup.
- Children can have mixed movement disorders, with stereotypies and tics or chorea, as demonstrated in the case vignettes.
- Referral to a pediatric movement disorder specialist can be useful when the type of movement is not straightforward and when a higher level of management (eg, with botulinum toxin or deep brain stimulation) is being considered.

jumping, repetitive neck extension, and eye rolling that occurred when he played video games. His movements did not cause educational or psychosocial issues, but they did cause physical impairment. There was a family history of tics. He had a normal neurologic examination, with few vocal sounds and motor movements. Conservative management was discussed with his parents.

DISCUSSION

Do Abnormal Movements Appear Similar in Children and Adults?

Children often present to pediatric and primary care clinics with unusual movements that can be challenging to describe, which contributes to uncertainty in their diagnosis, evaluation, and treatment. Movement disorders are characterized by exhibiting too much (hyperkinetic) or too little (hypokinetic) movement, and hyperkinetic movement disorders are more common in children.[3–5] Identification of childhood-onset movement disorders can be facilitated by the presence of additional cues, such as family history, the nature of the movements, medical history, and additional examination findings.

When evaluating hyperkinetic movement disorders, the first step is to determine if they are continuous or intermittent. If they are continuous, clinicians should assess whether the movements are slow and writhing (athetosis), dance-like (chorea), or of large amplitude (ballism). If they are intermittent, it is helpful to determine if the movements are brief (quick and jerky [myoclonus] or associated with a premonitory urge [tics] or if they can be redirected (eg, stereotypies). If they are prolonged, they can be categorized as rhythmic (tremor) or with a sustained posture (dystonia). Individuals can also have mixed movements (eg, choreoathetosis, dystonia, and myoclonus).

Early diagnosis can lead to effective management/treatment for treatable conditions.[4,6] Labeling the type of movement is the first step in evaluation of a child with unusual movements[4,7]; not all unusual movements in children are abnormal.[7] A careful movement history, a medical history (with associated neurologic/nonneurologic features and time course), a detailed physical examination, and a family history can provide clues to the nature of movement disorders.

It is also important to elicit from the child the impact that the movements are having on his/her quality of life, including but not limited to, his/her psychosocial, educational, and physical well-being.[4,5,7] Furthermore, during an evaluation it is useful to identify triggers, aggravating or relieving factors, preceding urges, suppressibility, and distractibility of the movements.[5] Unfortunately, the course of movement disorders can be difficult to predict: they may remain stable or shift in their manifestations, severity, and distribution over time.[8]

What Causes Abnormal Movements?

In adults, the etiology of hyperkinetic movements is most often exposure to neuroleptics, trauma of the brain and extremities, comorbid neurologic conditions (such as a prior stroke or Huntington's disease), mitochondrial disorder, or infectious diseases, which can be psychogenic or genetic as determined through family history, genetic testing, and/or ruling out other causes.[3,9,10] Psychogenic movements are considered when there is incongruency or distractibility from symptoms, regardless of psychiatric symptoms being present.[10]

However, the etiology for hyperkinetic movement disorders in children is most often attributed to metabolic conditions, infectious diseases, drugs/toxins, brain structure abnormalities, and hereditary/neurodegenerative conditions or is considered idiopathic when genetic testing and family history are negative.[8]

What Can Abnormal Movements Look like?

Since hyperkinetic movements are heterogeneous, an accurate description of the movements is crucial to understand the etiology.[3] It is worth noting that the etiology and prevalence of movement disorders in children may be different than in adults.[7] For example, tics are the most common movement disorder in pediatric clinics[4,6,7]; however, in adults (especially in those over the age of 50 years), the most common movement disorder is essential tremor followed by Parkinson disease.[3]

What Are Tics?

Tics tend to be quick, repetitive movements that are often preceded by a premonitory urge/sensation; they are usually partially suppressible. Moreover, tics tend to wax and wane, changing in complexity or presentation over time.[11] They exist on a neurodevelopmental spectrum with provisional (transient) tic disorder, chronic (persistent) tic disorders, and TS. Individuals meet diagnostic criteria for transient tic disorder when tics have been present for less than 1 year, whereas individuals meet criteria for chronic persistent tic disorder when vocal or motor tics have been present for longer than 1 year. Individuals meet diagnostic criteria for TS when multiple motor and vocal tics have been present for at least 1 year.[11] Tics occur disproportionally in males, with a 3:1 ratio in TS and a 2:1 ratio with chronic tic disorders.[1]

The mean age at onset for tics is between 4 and 6 years of age with tics worsening in adolescence; however, they may improve over time.[1,11] Only a small percentage of patients become more symptomatic and experience debilitating tics into adulthood.[1,11]

Those with TS are at risk for comorbid conditions, including attention-deficit/hyperactivity disorder (ADHD), which is present in 60% of those with TS, and anxiety/obsessive-compulsive disorders (OCD), which occur in 40%–60% of those with TS.[11] Since tics typically diminish in adulthood, adults with TS often experience the comorbid conditions, such as ADHD and anxiety/OCD as the most debilitating features.[1,11]

What Are Stereotypies?

Stereotypies are similar in phenomenology to tics, in that both exhibit repetitive movements.[5] However, in stereotypies there tends to be lack of premonitory sensations and the age at onset is much younger, with most individuals exhibiting symptoms before the age of 2 years.[5,12] There can be overlap between stereotypies and tics, and co-occurrence of these movement disorders is demonstrated in case 3.[12] Examples of stereotypies include body rocking, head nodding, hand wringing, finger tapping, and pacing. Stereotypies often improve in late adolescence or early adulthood.

What Is Chorea?

Chorea is a disorder of involuntary movements that appears random in presentation; it is commonly mistaken for tics.[4,5] The movements are less stereotyped,[5] and it lies on a spectrum with athetosis (slow writing movements) and ballism (large flailing movements of the limbs).[3,13] Chorea has many causes, with Sydenham chorea being the most common acquired chorea in children; drug-induced chorea is the most common cause in adults.[2–5] Huntington's disease is a well-known genetic form of chorea in adults,[2,3] and children with juvenile-onset Huntington's disease often present with an akinetic rigid syndrome.[2] Other genetic causes of chorea in children include disorders related to mutations in *ADCY5, NKX2.1*, and *PDE10A*.[14] Anti–*N*-methyl-D-aspartate receptor encephalitis also causes chorea.[5]

What Is Dystonia?

Dystonia is a contraction/co-contraction of agonist/antagonist muscles that causes twisting or abnormal postures or repetitive movements.[4,13] The location of dystonia, age at onset, associated features, and ethnic ancestry and family history are important variables to consider in the evaluation. The *geste antagoniste*, also known as a sensory trick, is an observed phenomenon in some dystonia cases whereby a voluntary maneuver temporarily reduces the severity of symptoms. In 1 study,[15] it was reported to be most frequently observed in focal cases of dystonia and was less likely to occur in segmental cases. In children, dystonia typically begins in the lower extremities and in 50% of cases spreads to the rest of the body, whereas in adults, dystonia tends to remain localized to the neck (cervical area) or eyes (blepharospasm).[3] Pediatric dystonia is often secondary to cerebral palsy, but primary genetic causes can also occur with mutations in the *TOR1A, THAP1,* and *GCH1* genes among others.[5,16] Additionally, there are new emerging monogenic complex dystonia-causing genes, such as *ADCY5* and *KMT2B*.[14] Adult-onset of generalized dystonia is most often attributed to drugs or an associated neurodegenerative disorder.[3] Children with dystonia may present to an emergency department with *status dystonicus* or with drug-induced dystonia.[5]

What Is Tremor?

Tremor is a rhythmic oscillation (back and forth) movement around a joint or axis.[3,4] Tremor can also affect the voice[5] and can be categorized as either rest, action, rubral, physiologic, or psychogenic in nature.[8] In adults, the most common movement disorder is essential tremor, and another prevalent disease in adults, Parkinson disease, also often involves tremor.[3] The most common causes of tremor in children are physiologic, structural brain disorders (eg, stroke); metabolic disorders; drugs/toxins; functional; or hereditary (juvenile Parkinson disease, Wilson's disease, familial essential tremor).[8] In children, acute-onset tremor is most likely related to a functional movement disorder.[5] Different tasks and postures can evoke tremor and help with description, such as a tremor at rest or one that is evident either when maintaining a posture against gravity (known as postural tremor) or when making a voluntary movement (known as kinetic or action tremor).[3,5] Similar to chorea, the history and examination can guide diagnostic workup and treatment of tremor.[5]

What Is Myoclonus?

Myoclonus is a hyperkinetic movement that presents with quick, shock-like jerks that have a faster frequency than chorea.[5] An example of a benign myoclonic jerk is when one is on the edge of sleep and experiences a hypnic jerk.[3] The movement is most commonly described by its body distribution and its site of origin (cortical, subcortical, or spinal), the latter of which can be studied further with an electroencephalogram.[3] Myoclonus is less likely due to a benign condition, and it should prompt a more emergent evaluation. Myoclonus can be seen in epilepsy as well as in rare conditions, such as opsoclonus myoclonus.[5] When myoclonus is observed in pediatric cases with dystonia or chorea, it should prompt genetic testing since there are known pathological mutations such as *SGCE, TOR1A, ADCY5,* and *KCTD17*.[16] When myoclonus is the only feature of a patient and there is no forthcoming etiology, it should be labeled essential myoclonus.[3]

What Can Our Cases Tell Us About Movement Disorders and Their Management?

Each of the 3 children in our case vignettes were seen in an outpatient clinic for evaluation of hyperkinetic movement disorders. In case 1, stereotypies were noted, and in case 3, TS and stereotypies were diagnosed. The second case revealed

that not all hyperkinetic movements in children are tics; this child had stereotypies and choreiform movements that had been present since infancy. These cases emphasize the importance of making an accurate diagnosis, as treatment (even if the approach taken is conservative) is predicated on etiology. If medications are considered later in life, thoughtful treatment can minimize drug side effects.

Discussion of Differential Diagnosis, Management, and Treatment

Our case vignettes of children and adolescents seen in an outpatient clinic for evaluation of hyperkinetic movement disorders can reveal an approach to diagnosis. Case 1 was a child with stereotypies, whose movement began at an early age (less than 2 years of age), lacked a premonitory urge, and did not change over time. Case 2 was a child with stereotypies and likely a genetic chorea. The stereotypies involved persistent movements of his head with extension of his arms and legs; they were predictable in nature. The choreiform movements had been present since infancy, and he also had signs of motor impersistence. Case 3 was a child with concurrent TS and stereotypies. Prior to onset of motor and vocal tics, he had predictable movements, such as jumping or hand flapping, that persisted and had not changed over time.

How Can Abnormal Movements Be Managed?

Evaluation, management, and treatment of hyperkinetic movement disorders are typically based on clinical history, time course, and neurologic examination.[3,4] Management of hyperkinetic movements typically involves use of a combination of medication and targeted physical therapy and may involve other treatment modalities, such as chemodenervation with botulinum toxin (eg, dystonia) and deep brain stimulation (eg, dystonia and some genetic forms of chorea). However, their etiology and aggravating factors should be considered to rule out other potential causes and to guide management. The specific movement disorder diagnosis will have important implications on the approach to management, even if only pharmacologic treatment is deemed necessary.

What Does the Assessment of Abnormal Movements Entail?

Assessment of abnormal movements requires a detailed history of the movements. If the movements are not present in the clinic, families should be asked to videotape the movements. The first steps include identifying the type of movement and eliciting other clues from the history and physical examination. For example, as in case 2, individuals with chorea may exhibit signs of motor impersistence. Workup in acute chorea includes brain imaging.[5] Additional metabolic and genetic testing should be performed based on history, examination, and family history.[5]

As in the first and third cases, management of tic disorders and stereotypies is geared toward treating the most impairing symptoms. Management of stereotypies often includes habit-reversal therapy.[12] In tic disorders, if symptoms are not bothersome, a conservative approach (with monitoring of tics and co-occurring conditions) is appropriate. Pharmacologic treatment in conjunction with other therapies (such as habit-reversal therapy and comprehensive behavioral intervention for tics) can be helpful.[11]

CONCLUSION

We provided a broad overview of hyperkinetic movement disorders in children, tips for proper evaluation, clues to creation of a differential diagnosis, and pointers for treatment and management. We also highlighted some differences in hyperkinetic movements between children and adults and emphasized the importance of gathering an accurate family history by drawing a family pedigree,[3] as it can provide clues to underlying diagnoses (such as is seen with dystonia, tics, stereotypies, myoclonus-dystonia, and chorea).[1,16]

In summary, when evaluating a patient with extra movements, it is important to identify what type of movement it is (eg, dystonia, chorea, tics, tremor), elucidate the time course (acute, subacute, chronic), any associated features, and family history. Individuals with acute chorea, tremor, and myoclonus may warrant further diagnostic workup, such as brain imaging. The diagnosis of individuals with tics/TS and stereotypies is mainly clinical in the setting of a normal neurologic examination. Be mindful that there can be overlapping features of movement disorders. To improve the quality of life, treatment should be tailored toward the underlying cause and the degree of impairment. In each of our cases, the parents preferred to embark upon conservative management with watchful waiting.

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